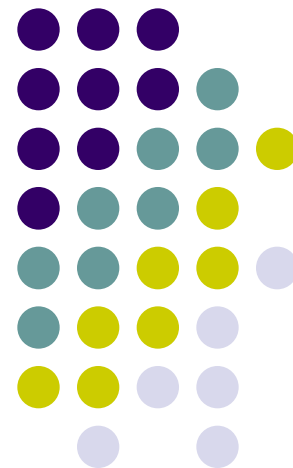


# Kidney & Other Autoimmune Diseases

**Hussein Sheashaa, MD**

Professor of Nephrology and manager of Quality Assurance Unit, Urology and Nephrology Center and Director of Medical E-Learning Unit, Mansoura University







President, Mansoura University  
**Prof. AL-Sayed Abdel**

Vice President, Mansoura University  
**Prof. Magda Nasr** Dean

Head of Internal Medicine  
**Prof. Megahed Abo EL**

Meeting Honorary President  
**Prof. Mohamed Sob**  
**Prof. Ali Badr**

Meeting President  
**Prof. Nagy Sayed Al**

Meeting Moderator  
**Dr. EL Shaat Ali**  
**Dr. Ahmed Abd EL-W**  
**Dr. Mostafa Abd EL-S**  
**Dr. Mohamed Wah**









Annual Meeting of Nephrology Unit  
Internal Medicine Department  
Mansoura University Hospitals  
16-18 April 2014  
Zakaria Nephrology Group (ONG)  
Al-Dakahlia Medical Syndicate

**Kidney in  
Systemic  
Diseases**

Under Patronage of  
**Prof. Al-Sayed Abdel Khalek**  
President, Mansoura University  
**Prof. Ehab Saad**  
Dean, Faculty of Medicine

Meeting  
Dr. Ahmed  
Dr. Mostafa

Prof. Nabil Lyoun

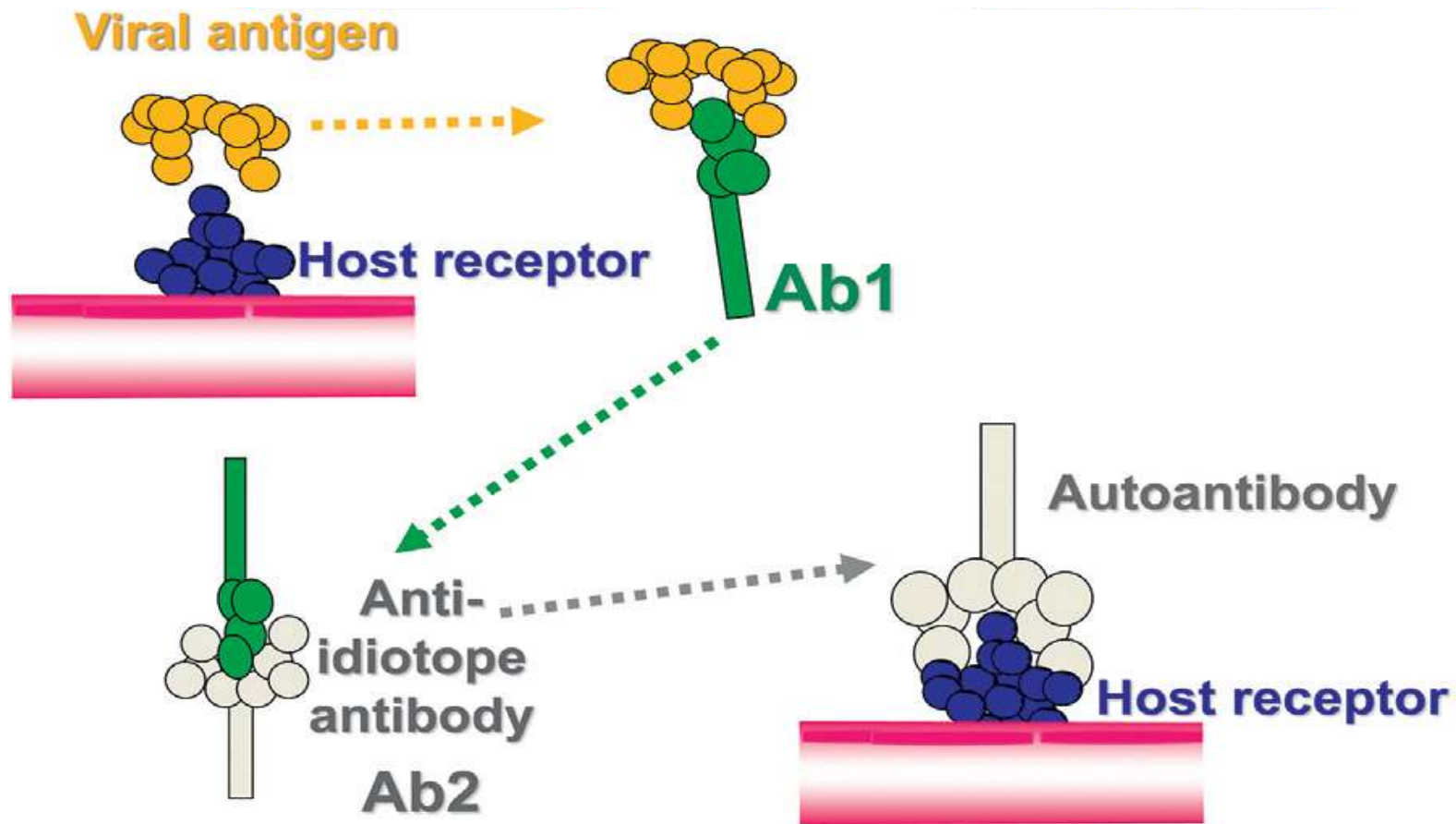
# Outline

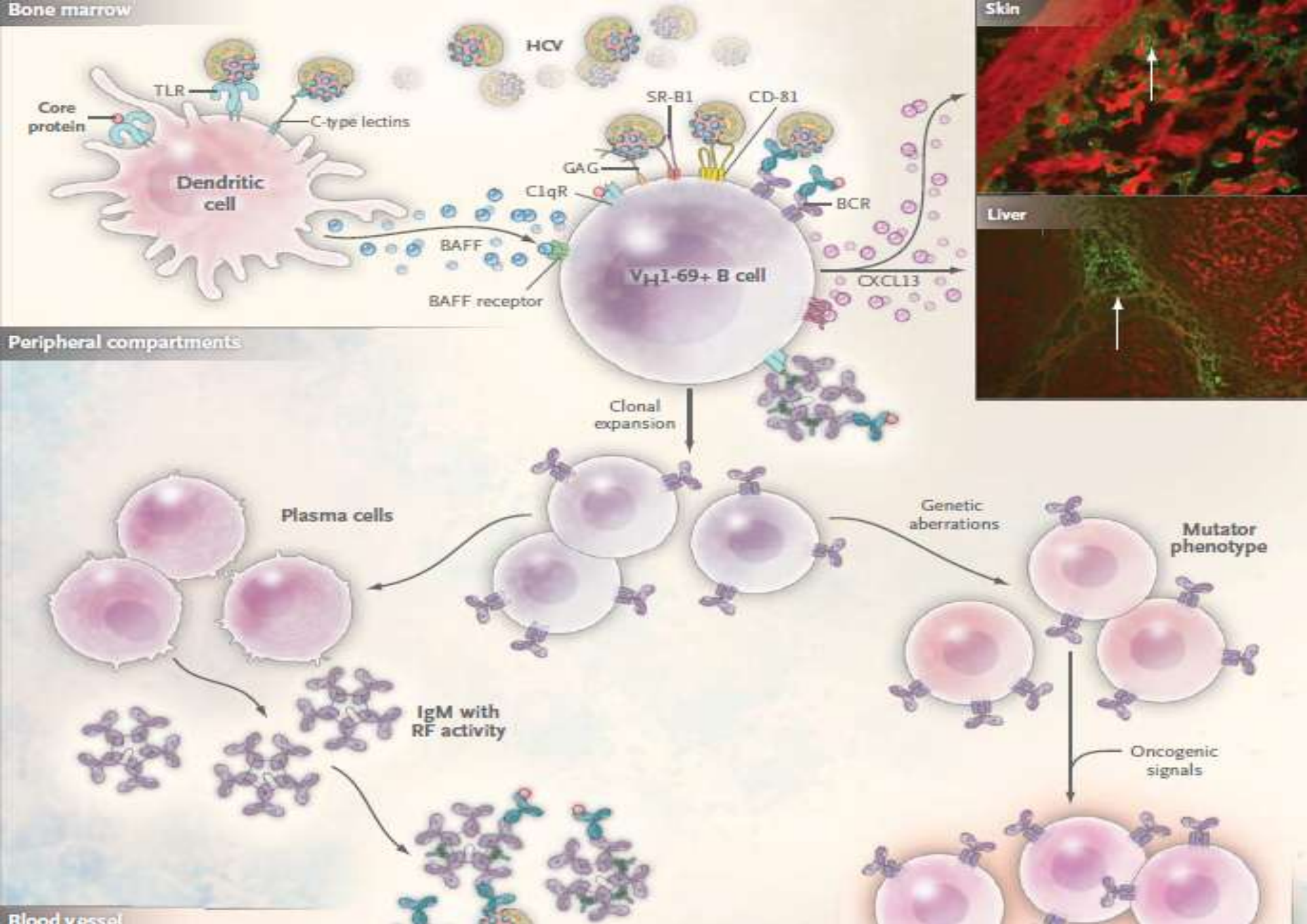


- ❖ **Introduction**
- ❖ **Kidney involvement in individual CT diseases**
- ❖ **APSN**
- ❖ **Miscellaneous autoimmune diseases**
- ❖ **Quiz**
- ❖ **Closure**

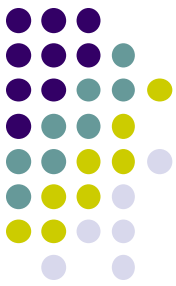


# Hallmarks of Autoimmune Diseases of The Kidney







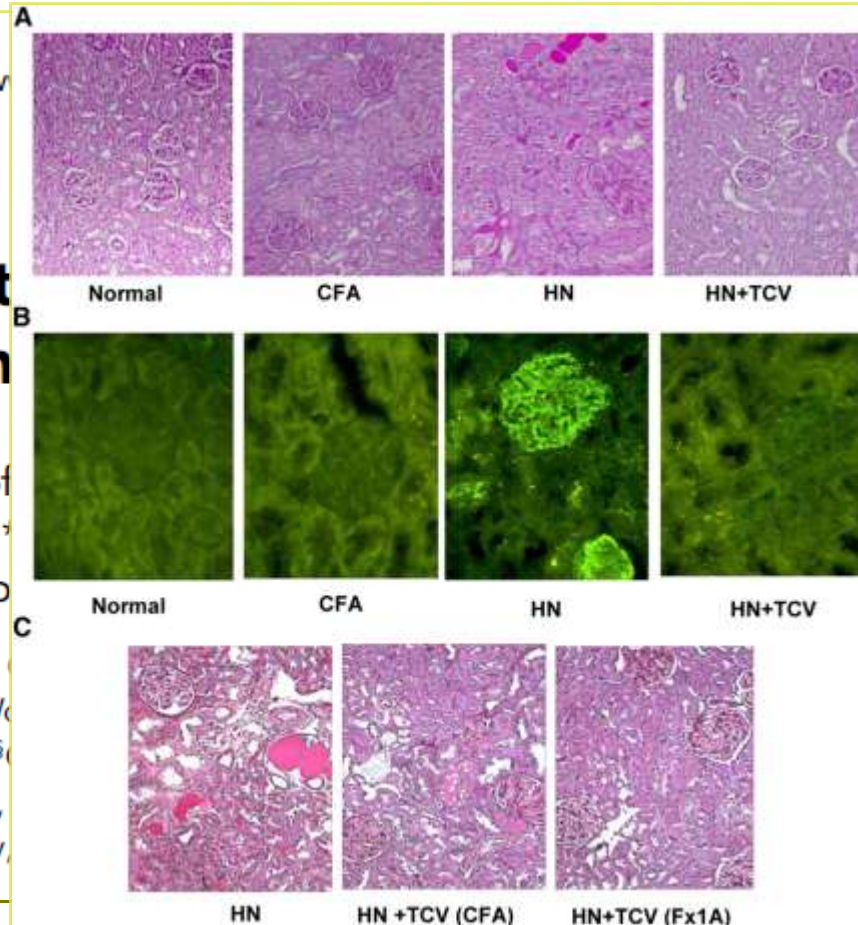


## BASIC RESEARCH

# CD8+ Regulate Protect Against

Yuan Min Wang,<sup>\*</sup> Geof  
Jimmy Jianheng Zhou,<sup>\*</sup>  
Xin Maggie Wang,<sup>||</sup> Yip

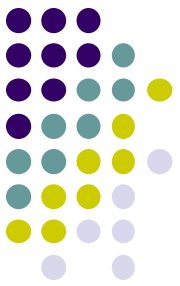
<sup>\*</sup>Centre for Kidney Research,  
University of Wollongong, Wollongong,  
Hospital, Sydney, Australia; <sup>§</sup>  
Millennium Institute, Sydney,  
University of Sydney, Sydney,



# I Vaccination

Sawyer,<sup>\*</sup>  
,<sup>‡</sup> Ya Wang,<sup>§</sup>  
Alexander<sup>\*</sup>

Centre for Medical Bioscience,  
Group, Royal Prince Alfred  
of Sydney at Westmead  
Millennium Institute,



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## Minireview

# B Cell Depletion: Rituximab in Glomerular Disease and Transplantation

S. Marinaki C. Skalioti J.N. Boletis

Department of Nephrology and Renal Transplant Unit, General Hospital 'Laiko', Athens, Greece

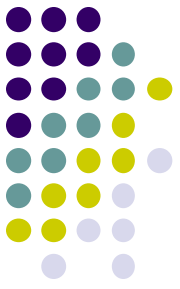


# CT Diseases

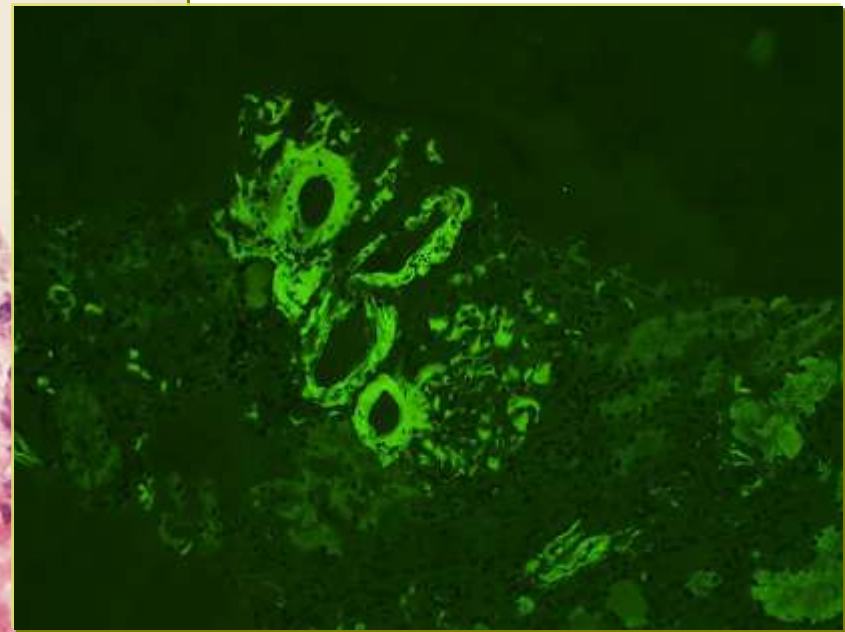
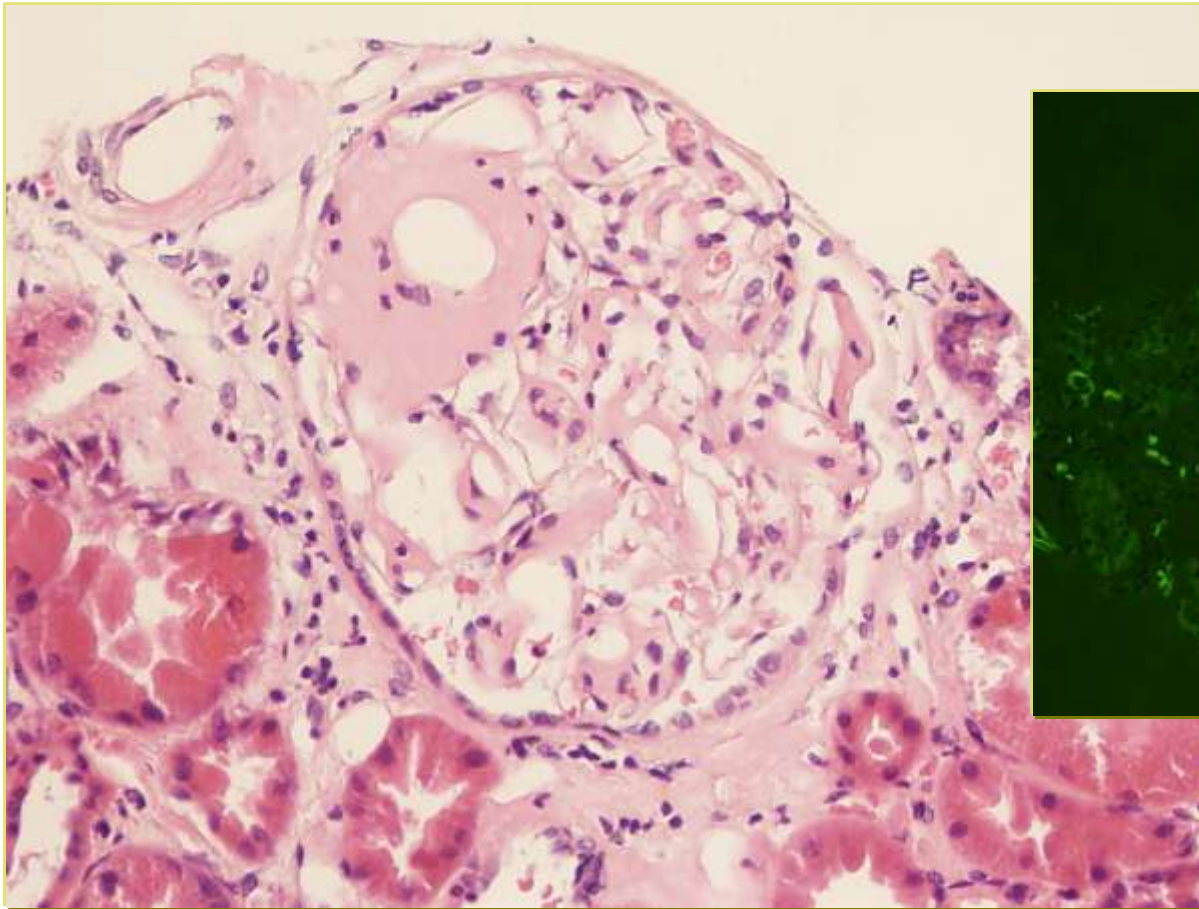
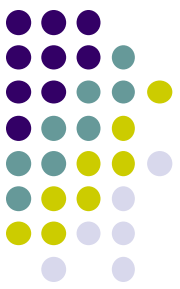


# Rheumatoid Arthritis:

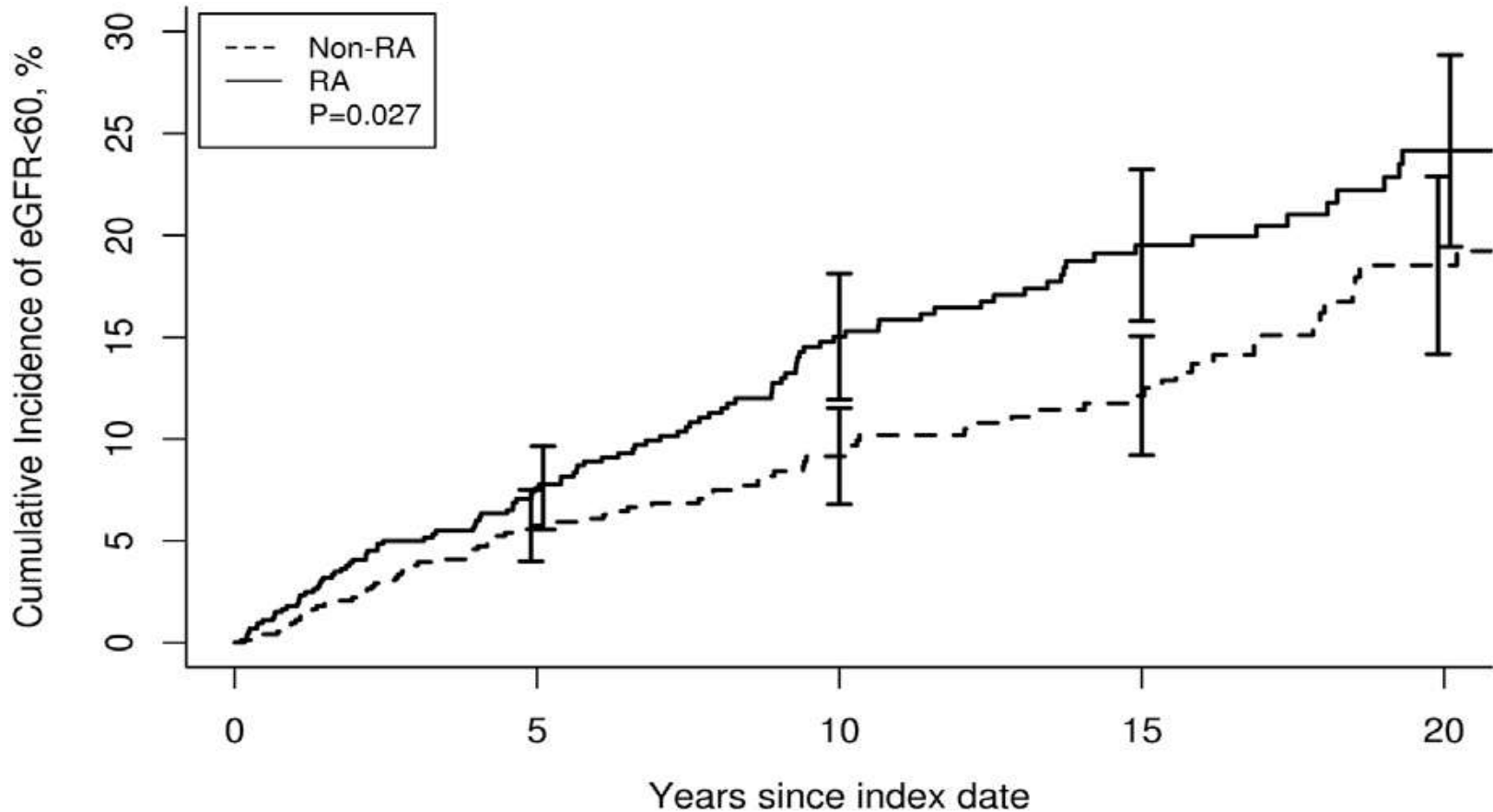
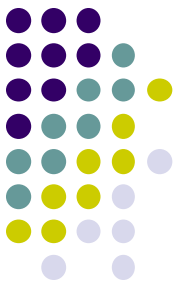
## Renal Biopsy Findings



- TIN
- Mesangioprolif. GN (IgA)
- FSGS
- MCD
- MN
- Amyloid
- Fibrillary GN
- Necrotizing cresc. GN
- Focal prolifer. GN

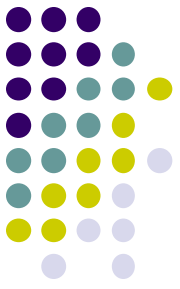


# Rheumatoid Arthritis: Reduced Kidney Function





# Sjögren syndrome: Renal Biopsy Findings



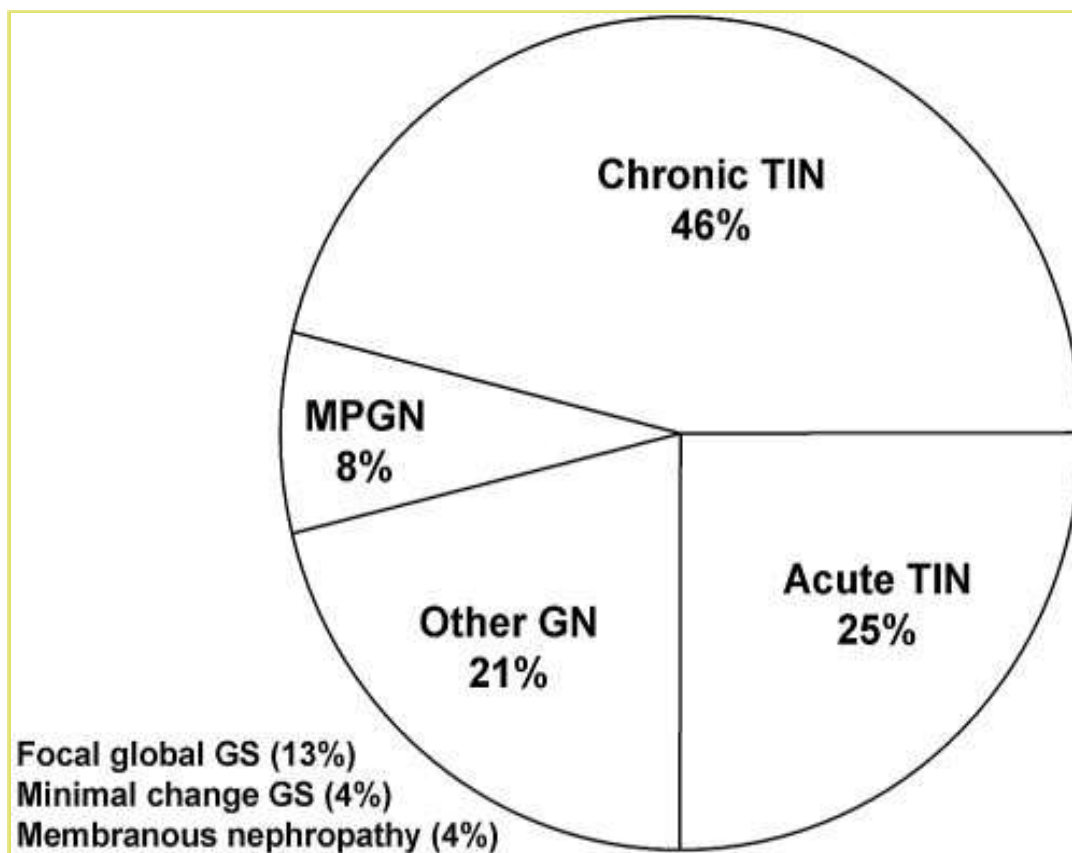
- TIN
- Mesangioprolif. GN (IgA)
- FSGS
- Cryog. MPGN
- MCD
- MN
- Amyloid

# Sjögren syndrome: Renal Biopsy Findings



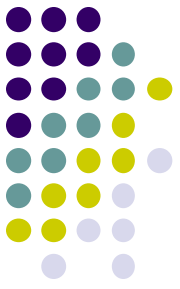
Patient #	Gender	Number of CC	Year of Biopsy (age in yr)	Clinical Presentation	Proteinuria* at Biopsy	Cr $\pm$ (MDRD) at Biopsy	Renal Biopsy Findings <sup>b</sup>	Postbiopsy Treatment	Follow-Up (mo) and Complications	Cr $\pm$ (MDRD) at Follow-Up
1	F	3	2001 (63)	CKD	<0.3	1.7 (32)	FGGS	None initially, then azathioprine (2003), then mycophenolate (after 2004)	75	1.7 (32)
2	F	3	2003 (77)	ESRD, proteinuria	2.3	5.0 (9)	FSGS, mild TIN	P taper	4	3.2 (15)
3	F	4	1992 (58)	ARF, cryo (type I), HCV not done, low complement	6.5	3.5 (14)	MPGN, cryos, immune complexes	P taper	<1	2.4 (22)
4	F	3	1995 (47)	ARF, hematuria, cryo (type I), HCV negative, low complement	1.3	1.4 (43)	MPGN, cryos, immune complexes	P taper, HC, rituximab (2006)	148 Lymphoma	0.8 (78)
5	F	3	2004 (67)	CKD	<0.3	2.4 (21)	Global glomerulosclerosis, mild TIN, arteriosclerosis	P taper	17	2.2 (24)
6	F	3	1995 (66)	NR proteinuria	5.9	1.3 (44)	Minimal change, arteriosclerosis	P, HC	139	0.9 (65)
7	F	4	1978 (71)	ARF	0.6	1.3 (43)	Membranous nephropathy, immune complexes	None	<1	Unknown
8	F	4	2001 (45)	CKD, distal RTA	<0.3	1.7 (35)	TIN	P taper to 5 mg chronically	80	1.3 (46)
9	F	4	2001 (56)	CKD	0.38	1.4 (41)	TIN	P taper	60	1.4 (41)
10	F	3	2001 (70)	CKD	0.46	2.6 (19)	TIN	P taper, C (2002-2003), mycophenolate (2006-)	67	2.2 (23)
11	F	3	2005 (48)	CKD, distal RTA	0.76	1.8 (32)	TIN	P taper, rituximab	25	1.4 (42)
12	F	2 <sup>c</sup>	1998 (67)	CKD	2.3	1.6 (34)	TIN, FSGS	P taper, HC	96	2.5 (21)
13	M	3	1980 (66)	ESRD/HD, cryo (Type II) HCV status unknown.	1.2	8.4 (5)	TIN	P taper, plasma exchange	46 Lymphoma	5.0 (9)
14	F	4	1967 (58)	Fanconi syndrome, proximal RTA	2.2	1.9 (29)	TIN	P taper, C	192	1.8 (29)
15	M	3	1991 (49)	ARF, distal RTA	<0.3	2.8 (26)	TIN	P taper	3	1.2 (68)
16	M	3	1997 (57)	ARF	0.5	2.0 (37)	TIN	P taper, HC	<1	1.3 (59)
17	F	4	1996 (50)	CKD, cryo (Type II) HCV negative	0.8	2.1 (26)	TIN	P taper	38 Lymphoma	1.5 (39)
18	F	4	1990 (28)	Hypokalemia, distal RTA	1.5	1.9 (33)	TIN	P taper, HC	89	2.6 (22)
19	F	3	1991 (84)	ARF	<0.3	2.5 (20)	TIN	P taper	5	1.6 (33)
20	F	3	1998 (56)	ARF, history of sarcoidosis	<0.3	2.0 (27)	Granulomatous TIN	P taper, HC, rituximab (2006)	104 Lymphoma	1.2 (48)
21	F	4	1969 (36)	Distal RTA	1.2	1.3 (49)	TIN	P taper	<1	Unknown
22	F	2 <sup>c</sup>	1998 (15)	Distal RTA	0.47	1.1 (69)	TIN	P taper	28	1.1 (69)
23	F	4	1971 (65)	Distal RTA	<0.3	1.1 (53)	TIN	Plasma exchange	120	1.1 (52)
24	F	4	1982 (76)	CKD	<0.3	1.6 (33)	Arteriosclerosis, TIN	None	<1	1.9 (27)

# Sjögren syndrome: Renal Biopsy Findings

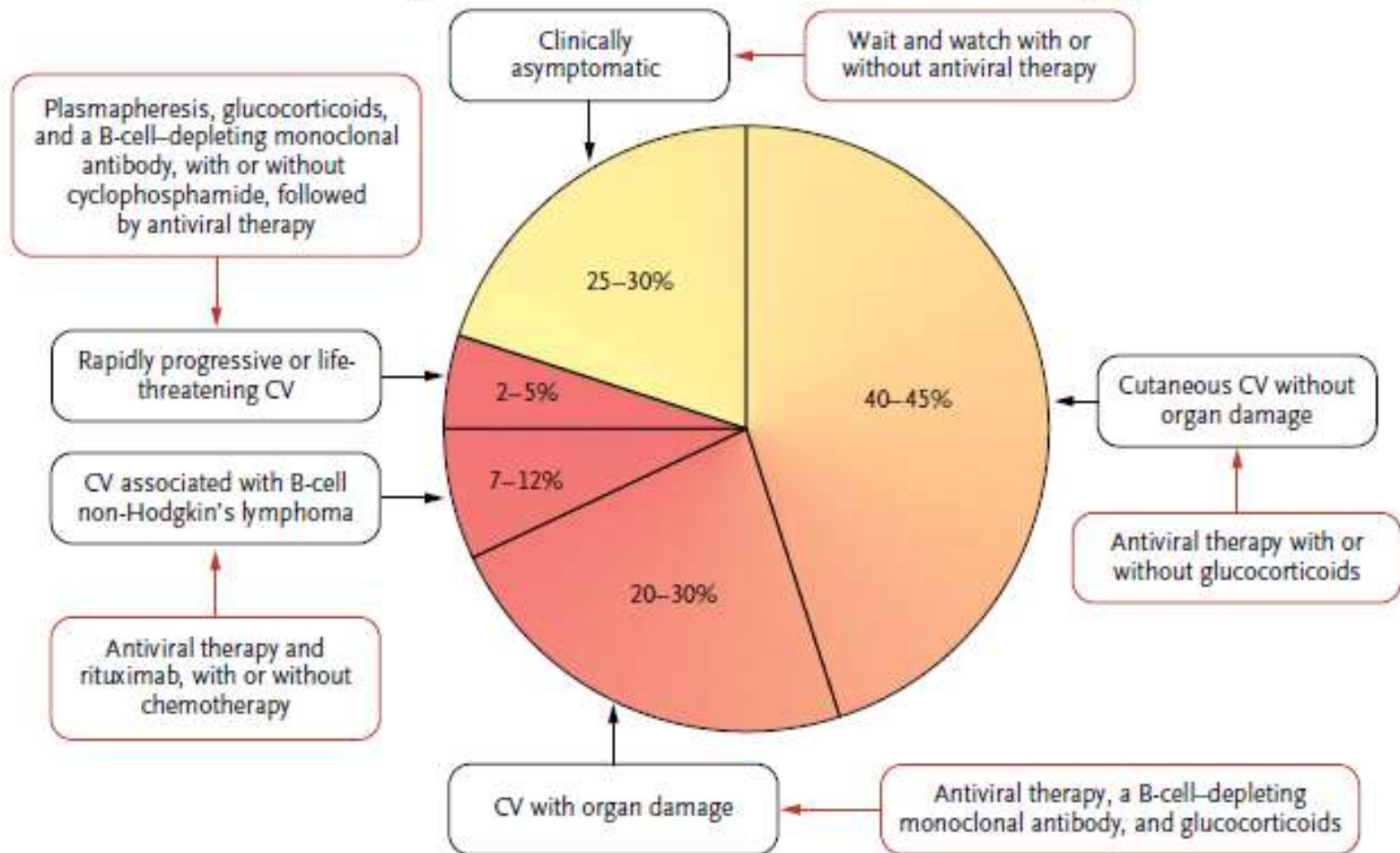
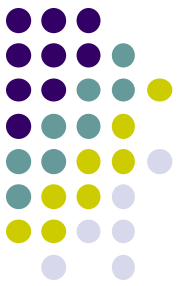




# Sjögren syndrome: Renal Biopsy Findings



# HCV-Related CV



# Polymyositis:

## Renal Biopsy Findings



- Mesangioprolif. GN (IgA)
- MCD
- MN
- Cresc. GN with FSGS

# Dermatomyositis:

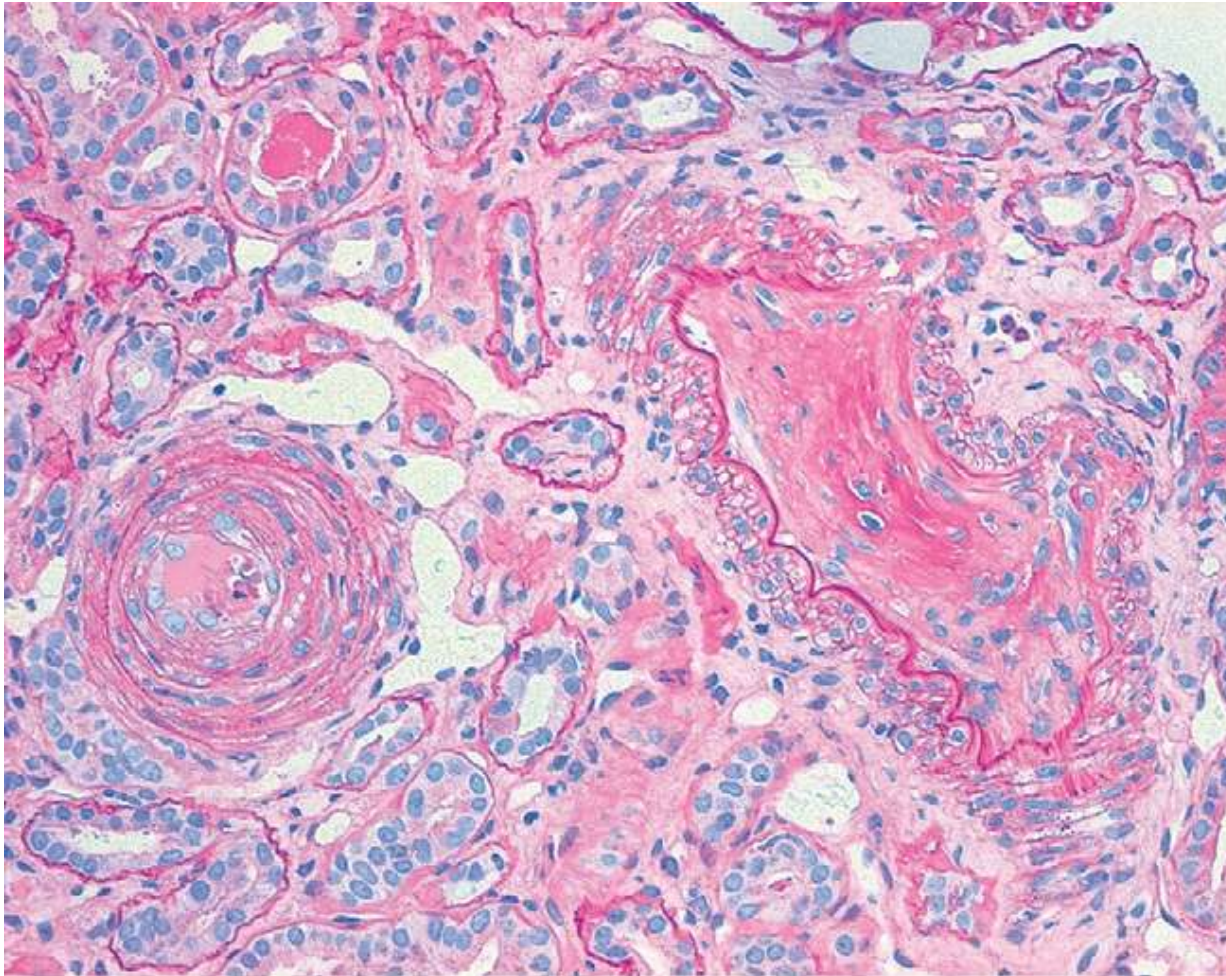
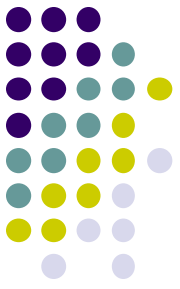
## Renal Biopsy Findings



- Mesangioprolif. GN (IgA)
- MN
- Diffuse proliferative GN



# Scleroderma Renal Crisis: Renal Biopsy Findings



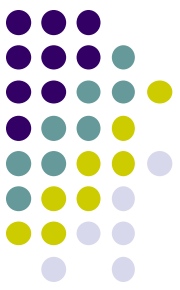
# Scleroderma Renal Crisis: Renal Biopsy Findings



- TMA
- Necrotizing GN

# Scleroderma Renal Crisis:

## Renal Biopsy Findings



### Findings Suggestive of TTP Rather Than SRC in Scleroderma

---

Severe thrombocytopenia (especially  $< 50,000/\mu\text{L}$ ) with MAHA

Hemorrhagic manifestations including purpura

Fever

Normal blood pressure

Treatment failure of ACE inhibitors

Severe deficiency of ADAMTS-13 activity with anti-ADAMTS13 antibody

---

TTP: Thrombotic thrombocytopenic purpura, MAHA: microangiopathic hemolytic anemia, SRC: scleroderma renal crisis, ACE: angiotensin converting enzyme

# Mixed CT Disease: Renal Biopsy Findings

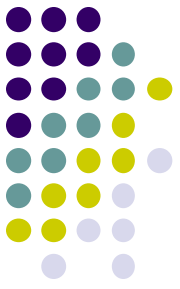


- MN or mesangial proliferative GN
- Diffuse proliferative



# Autoimmune Diseases:

## Renal Biopsy Indications



- rapid deterioration of renal function (exclude post renal and pre renal disorders first)
- proteinuria >1 g/d
- nephritic urine sediment with deterioration of kidney function
- increase in proteinuria/serum creatinine despite ongoing immunosuppressive therapy
- suspected interstitial nephritis
- diagnostic approach in case of uncertainties



# Antiphospholipid Syndrome

# Antiphospholipid Syndrome: Renal Biopsy Findings



- FSGS
- MCD
- MN
- TMA
- Fibrillary GN
- C3 Nephropathy

Volume 13 • Number 1 • January 2014

*NephSAP*<sup>®</sup>  
Nephrology Self-Assessment Program

Vaso-Occlusive Disorders  
and Kidney Disease

*Guest Co-Editors:*

*Patrick H. Nachman, MD*

*William F. Clark, MD*

*Vimal Derebail, MD*

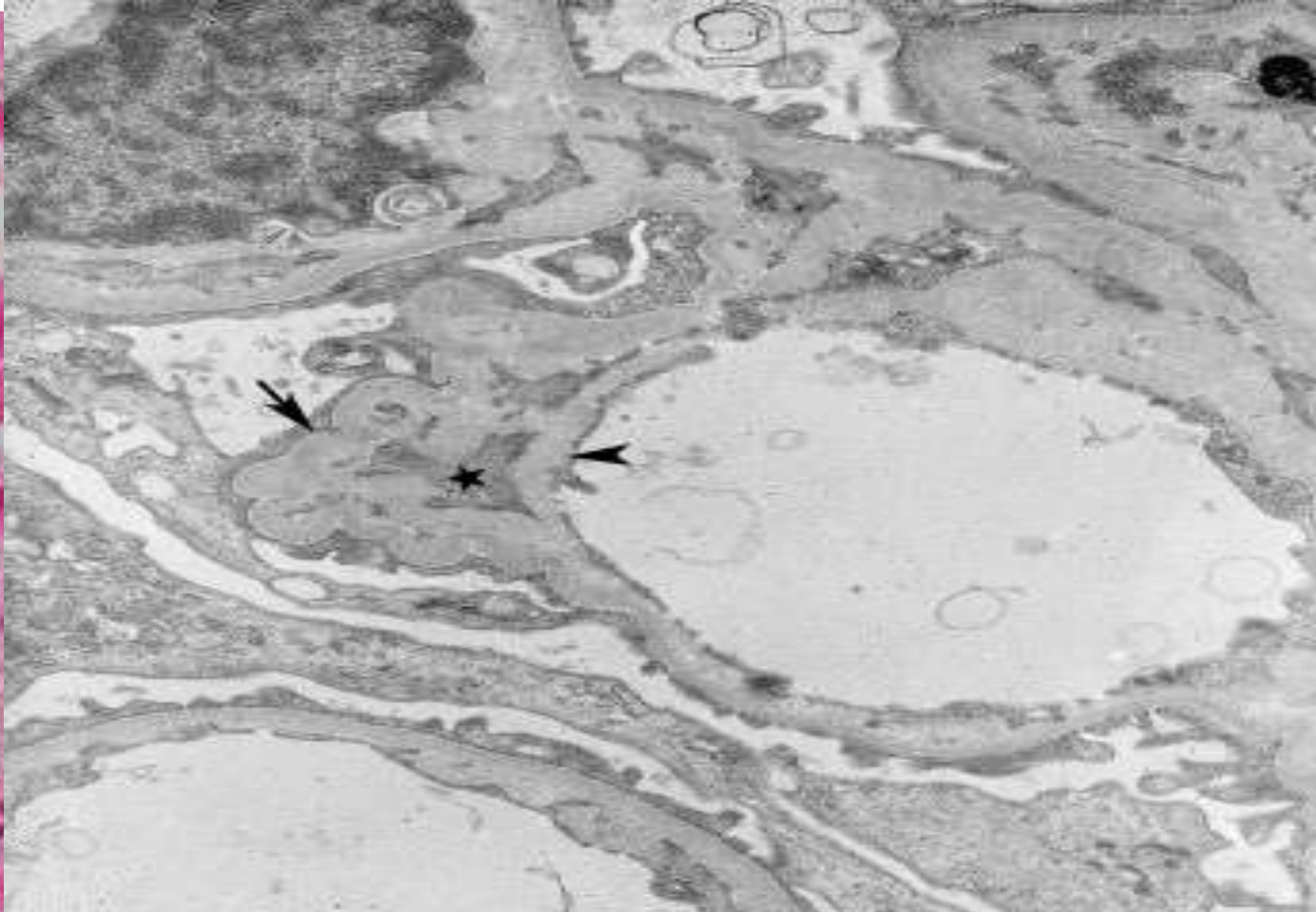
Nephrology Self-Assessment Program - Vol 13, No 1, January 2014

## Editorial

Antiphospholipid Syndrome Nephropathy: An Insidious Cause of Progressive Renal Failure

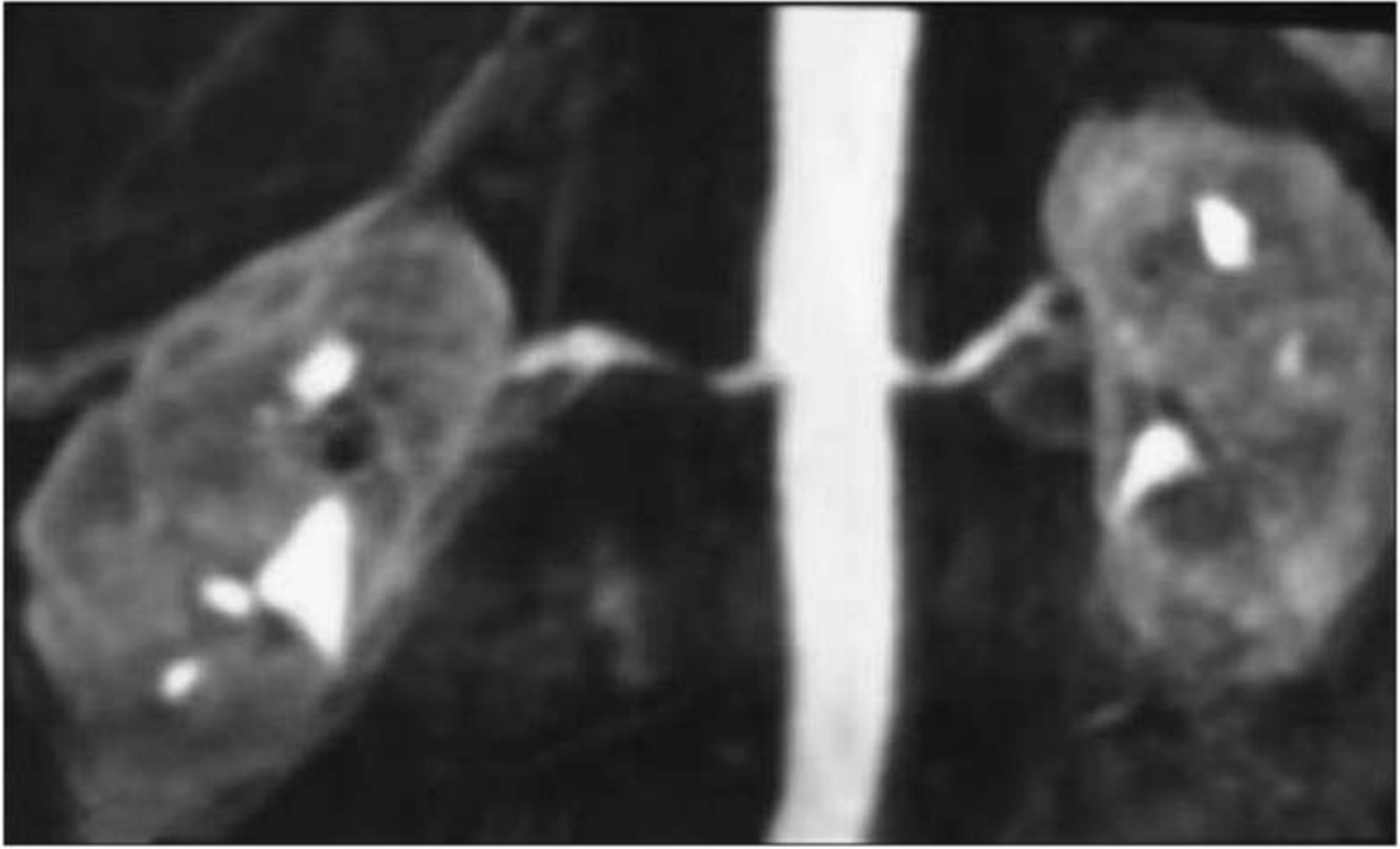
*Samir V. Parikh, MD, and Brad H. Rovin, MD, FASN, FACP*  
*Division of Nephrology, Wexner Medical Center, Ohio State University, Columbus, Ohio*

# APSN





# APSN



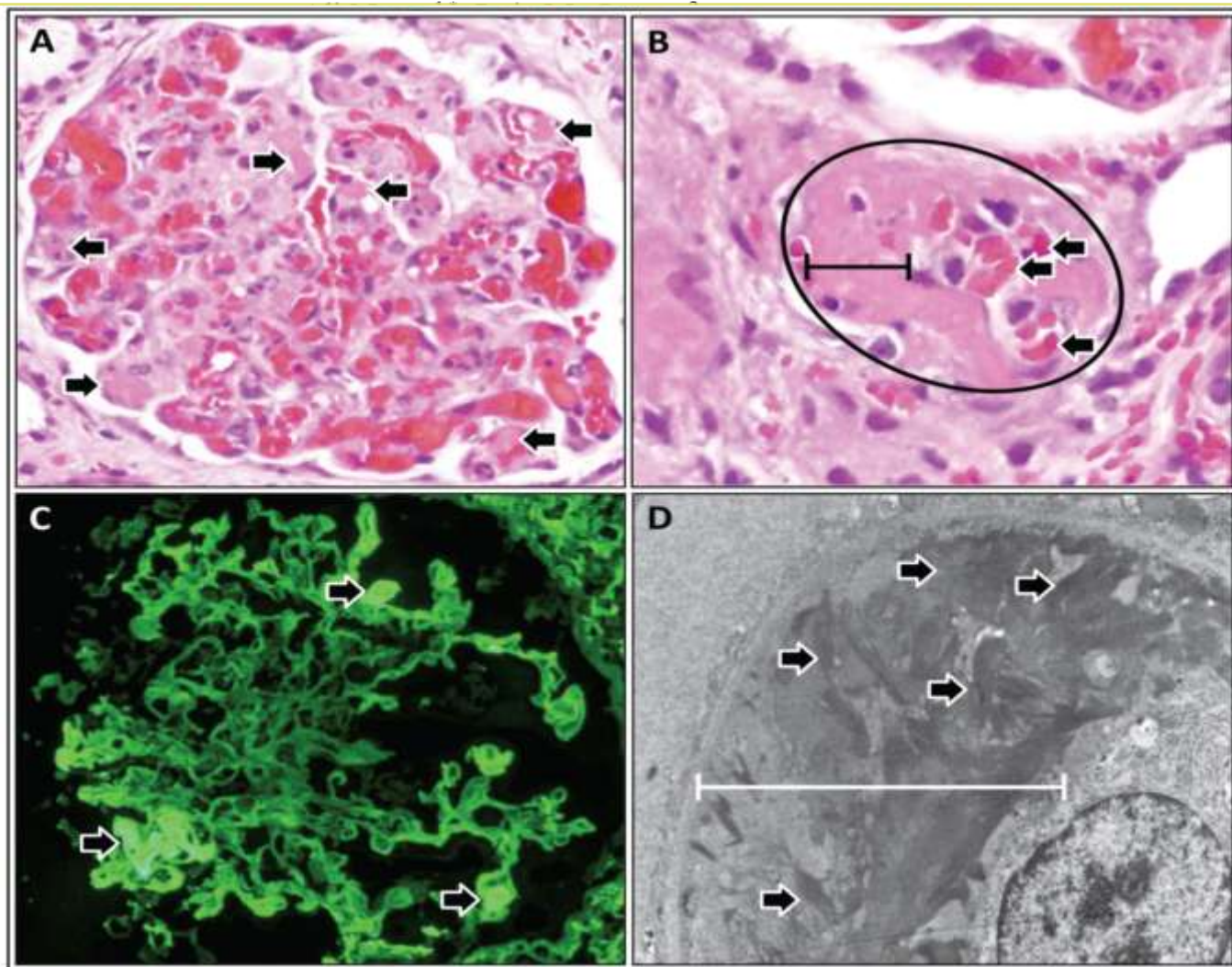
Nephrol Dial Transplant (2010) 25: 3147–3154

# Journal of Nephropathology

## Catastrophic antiphospholipid syndrome: a clinical review

<sup>1</sup>Division of Nephrology

<sup>2</sup>Division of Nephrology





# Miscellaneous



# Antiglomerular Basement Membrane Disease and Goodpasture's Disease

Richard G. Phelps, A. Neil Turner

## Factors Influencing Decision to Treat or Not to Treat Aggressively in Goodpasture's Disease

	Factors Favoring Aggressive Treatment	Factors Against Aggressive Treatment
Pulmonary hemorrhage	Present	Absent
Oliguria	Absent	Present
Creatinine	<5.5 mg/dl (approximately 500 $\mu$ mol/l)	>5.5–6.5 mg/dl (approximately 500–600 $\mu$ mol/l) and ANCA negative Severe damage on kidney biopsy No desire for early kidney transplantation
Other factors	Creatinine >5.5–6.5 mg/dl (approximately 500–600 $\mu$ mol/l) <i>but</i> Rapid and recent progression ANCA positive Glomerular damage less severe than expected Crescents recent, nonfibrous Early renal transplantation desired	
Associated disease	Absent	Unusually high risk from immunosuppression



# Atypical HUS



## *Etiology advanced*

- 1 Infection induced
  - a Stx-producing bacteria; enterohemorrhagic *E. coli*, *S. dysenteriae* type 1, *Citrobacter*
  - b *S. pneumoniae*, neuraminidase, and T antigen exposure
  - c Other infectious agents
- 2 Disorders of complement regulation
  - a Genetic disorders of complement regulation
  - b Acquired disorders of complement regulation, for example anti-CEH antibodies
- 3 Von Willebrand proteinase, ADAMTS13 deficiency
  - a Genetic disorders of ADAMTS13
  - b Acquired von Willebrand proteinase deficiency: autoimmune, drug induced
- 4 Defective cobalamine metabolism
- 5 Drug induced (quinine)

**Clinical  
Practice**

### Minireview

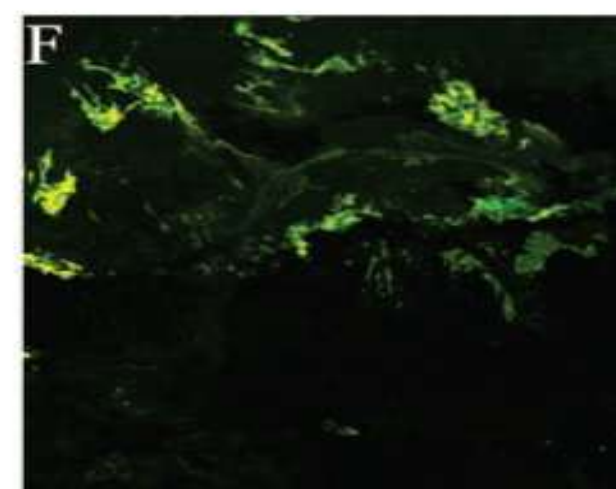
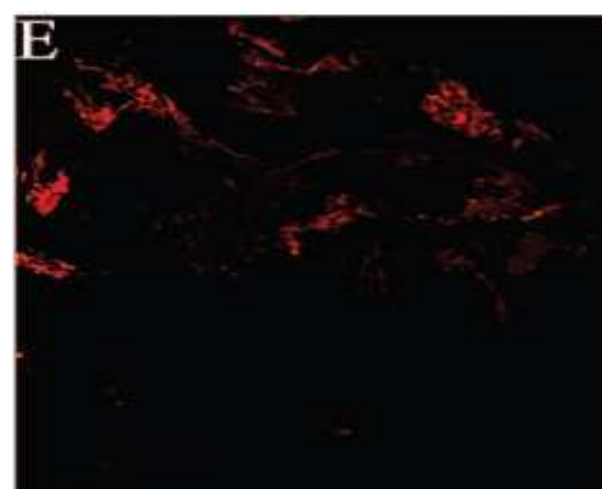
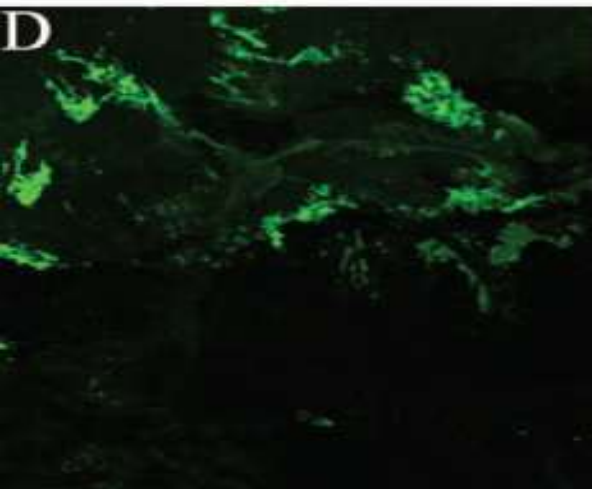
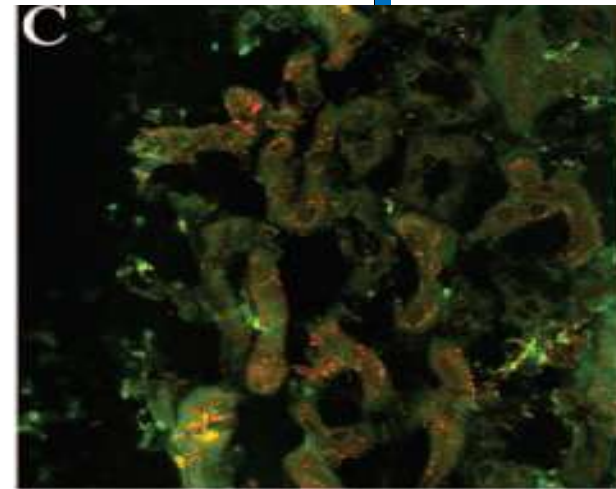
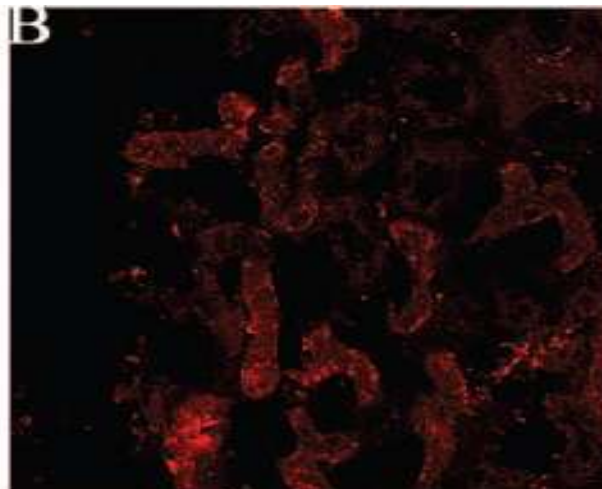
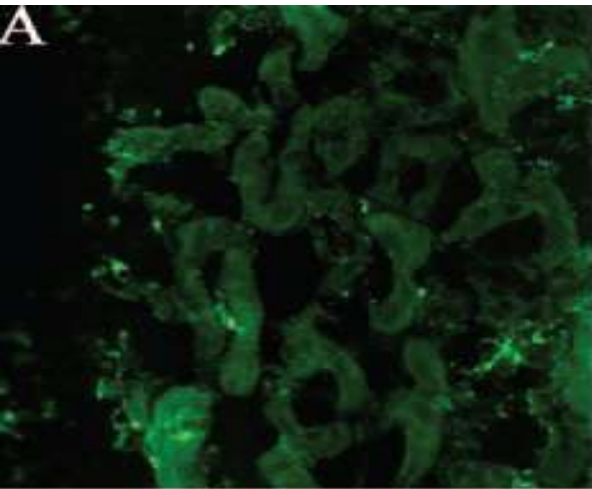
Nephron Clin Pract 2010;114:c219–c235  
DOI: 10.1159/000270545

## **Atypical Hemolytic Uremic Syndrome: Update on the Complement System and What Is New**

Patricia Hirt-Minkowski<sup>a</sup> Michael Dickenmann<sup>a</sup> Jürg A. Schifferli<sup>b</sup>

Divisions of <sup>a</sup>Transplantation Immunology and Nephrology, and <sup>b</sup>Internal Medicine, University Hospital Basel, Basel, Switzerland

# TINU Syndrome



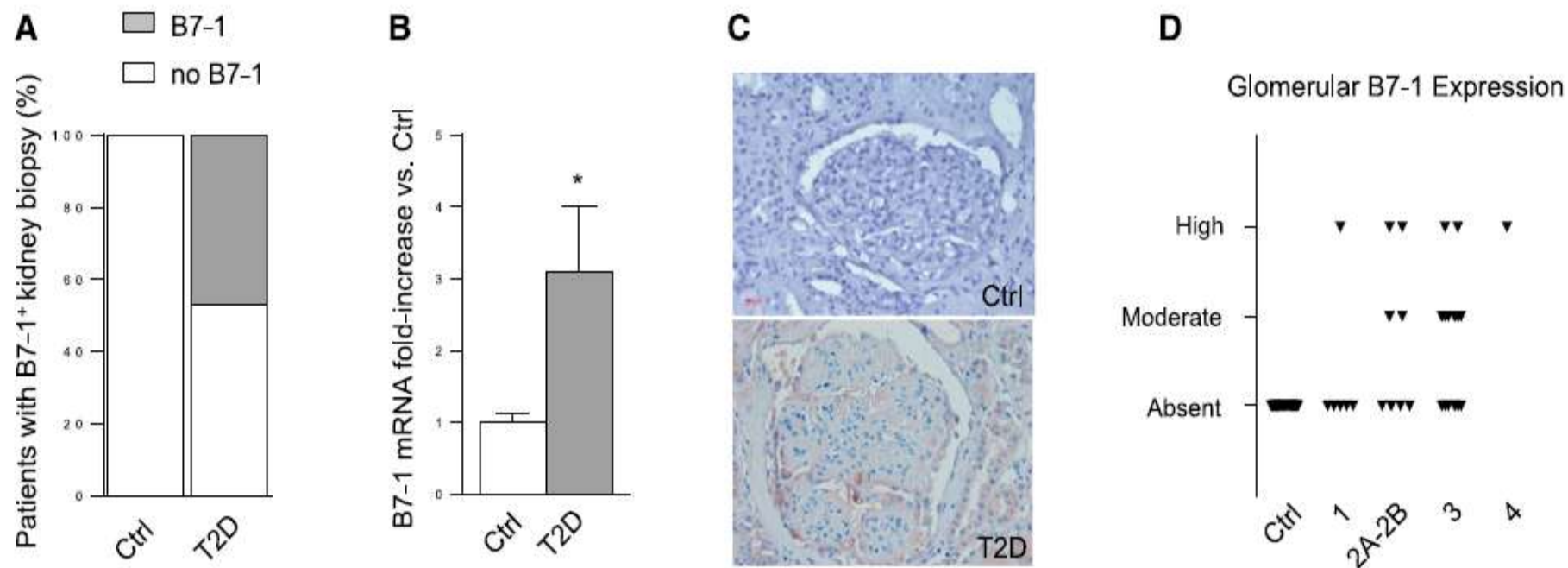
# Type 2 Diabetes



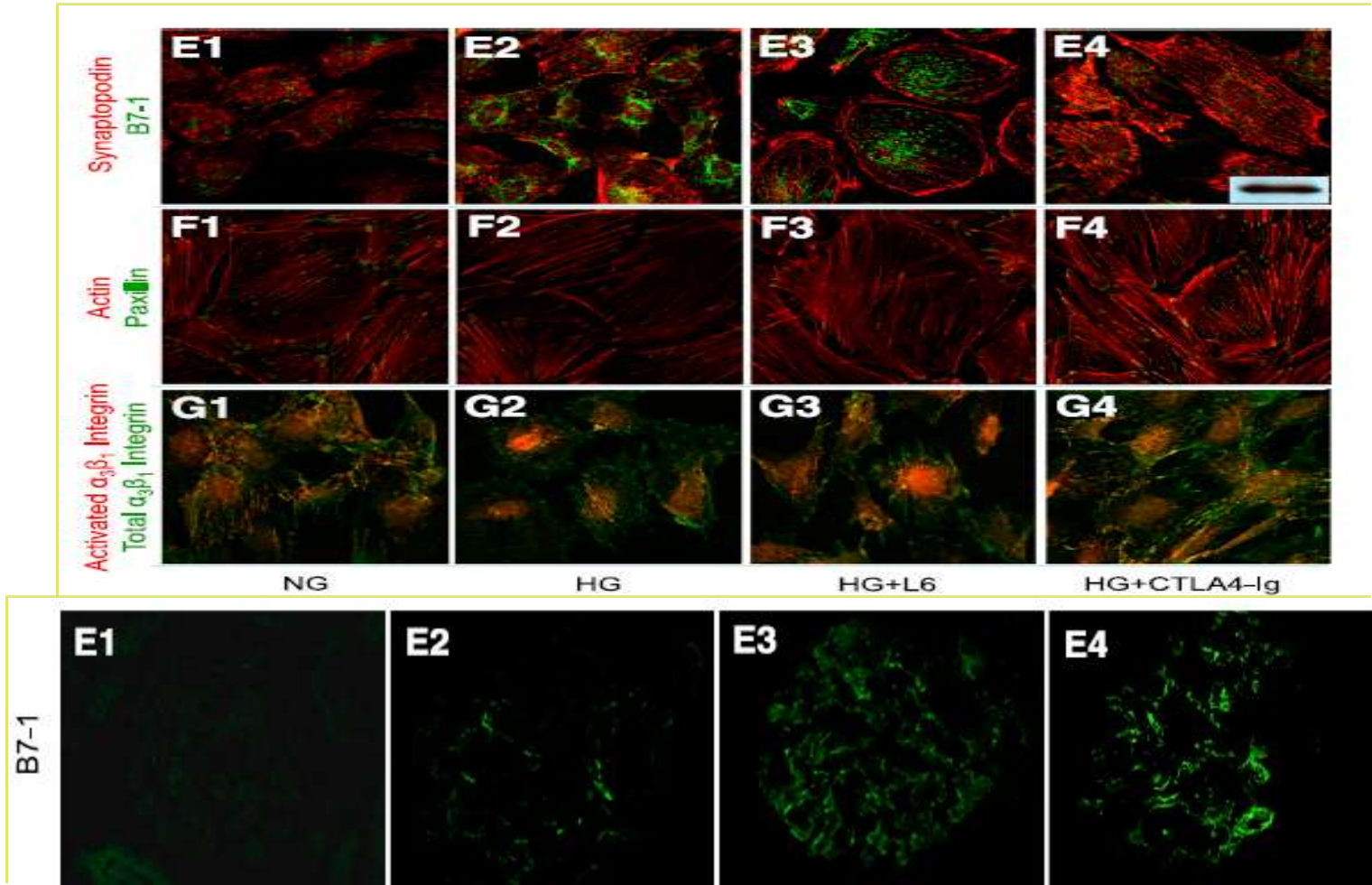
BASIC RESEARCH

www.jasn.org

## Role of Podocyte B7-1 in Diabetic Nephropathy



# Type 2 Diabetes

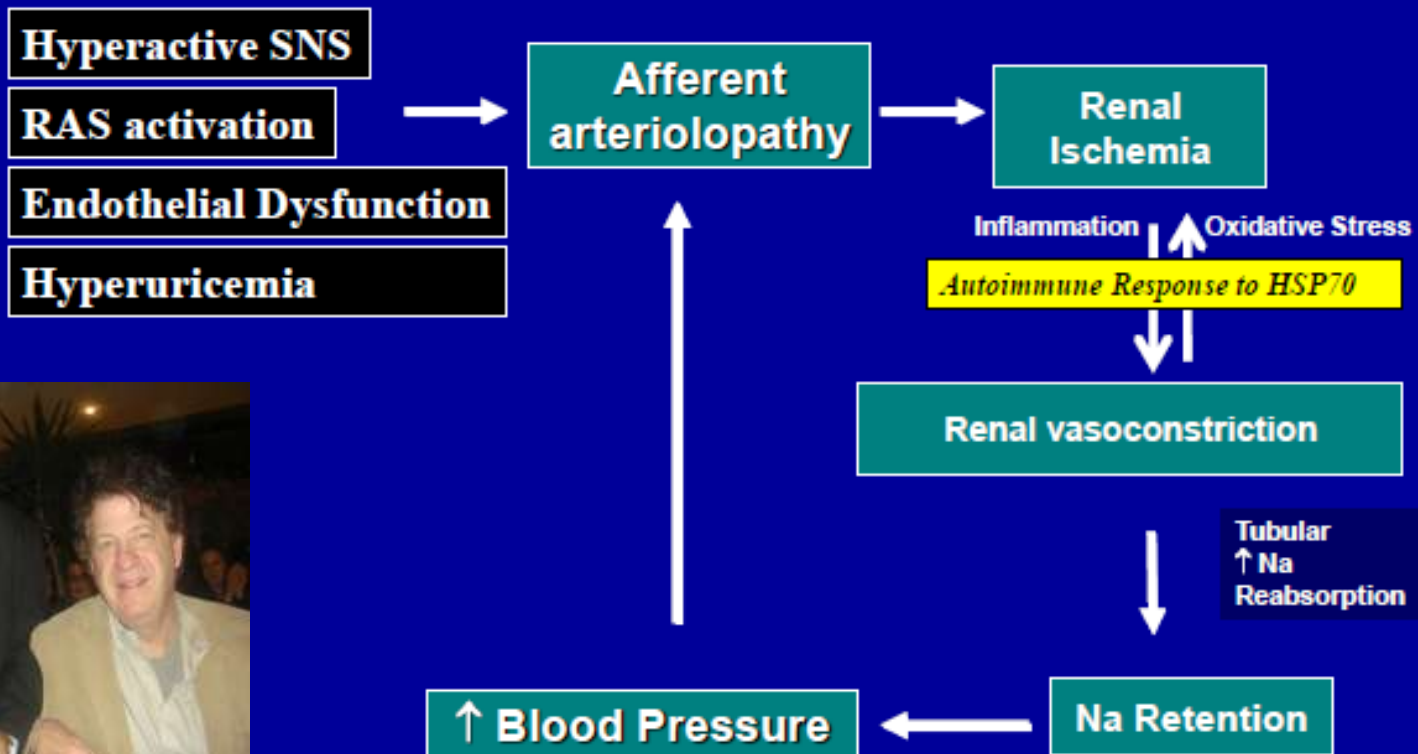




# A Role for T cells and Autoimmunity in Primary Hypertension?

**Richard J Johnson MD**  
**University of Colorado**

## A Pathway for Sodium Sensitive Hypertension





# GBS



## Case Report

nephron  
**Clinical  
Practice**

Nephron Clin Pract 2013;124:239–242  
DOI: [10.1159/000358087](https://doi.org/10.1159/000358087)

Published online: February 4, 2014

## Case Report: Guillain-Barré Syndrome following Renal Transplantation – A Diagnostic Dilemma

Adam D. Jakes<sup>a</sup> Poonam Jani<sup>b</sup> Sunil Bhandari<sup>b,c</sup>

<sup>a</sup>Leeds Teaching Hospitals NHS Trust, Leeds, <sup>b</sup>Hull York Medical School, University of Hull, and <sup>c</sup>Renal Unit, Hull and East Yorkshire Hospitals NHS Trust, Kingston upon Hull, UK



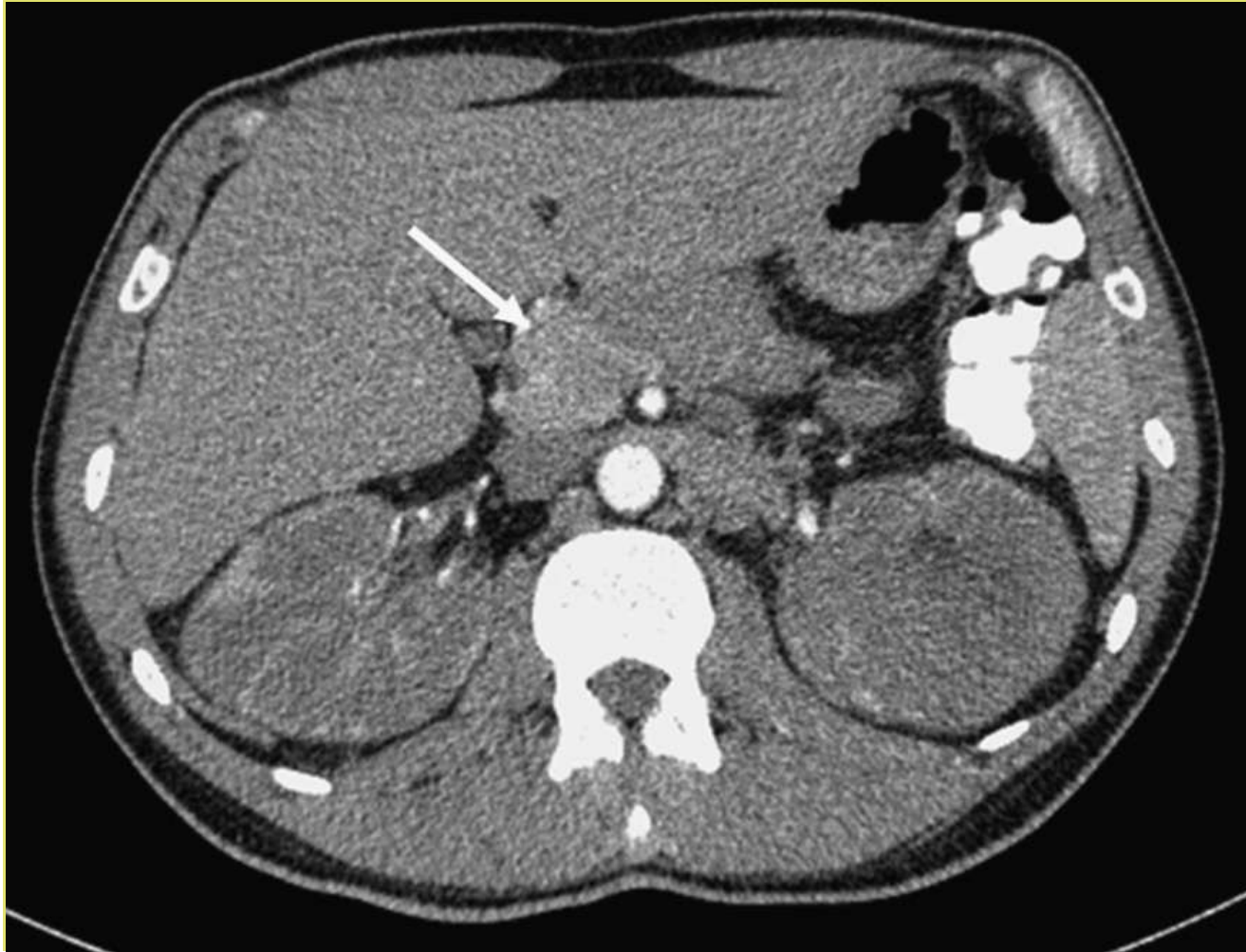
# Quiz and Awards

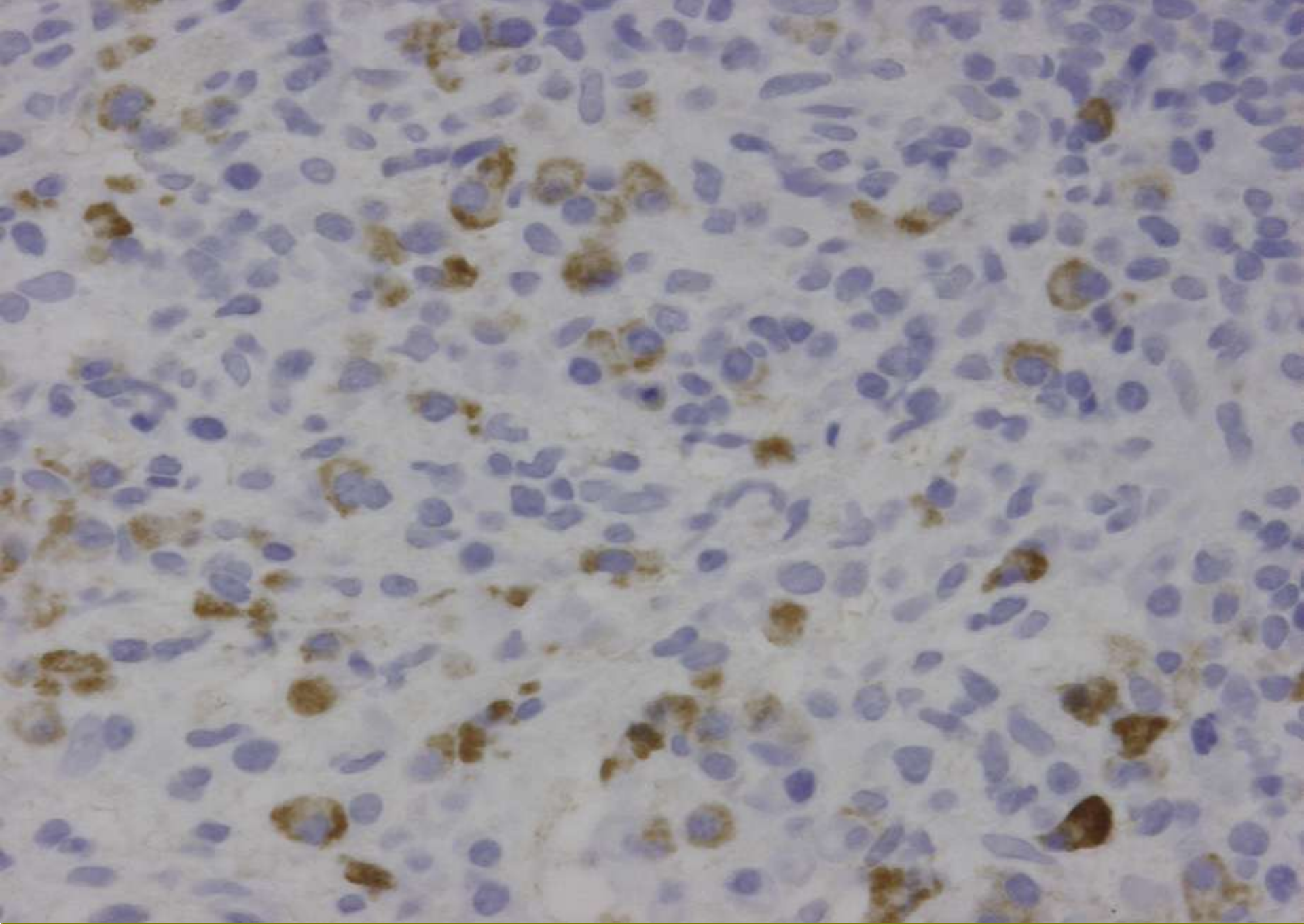
# Case Scenario



- A 46-year- old man
- Presentation:
  - ❖ Progressive weakness, arthralgia, hypertension
  - ❖ Weight loss of 4 kg
  - ❖ Proteinuria 1 gm/d
  - ❖ Increased serum creatinine from 1.4 to 2.6 mg/dl
  - ❖ Increased transaminases, serum albumin 3.1 and total protein 8.5

# Case Scenario

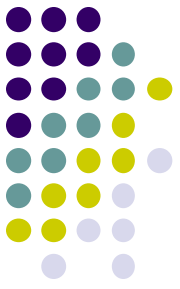






# **IgG4-Related Disease:**

## **Japanese Diagnostic Criteria**



### **Japanese CCD criteria \***

**1-Clinically diffuse / localised swelling**

**2-Elevated IgG4 > 135 mg / dl**

**3-Histopathology**

**-marked lympho & plasma cell + fibrosis**

**-infiltration by IgG4 + plasma cells \*\***

**Definite: 1+2+3, Probable: 1+3 Poss: 1+2**

**\*Japanese comprehensive clinical diagnostic criteria**

**\*\* IgG4+/ IgG+ >40% & 10 IgG4 + cells /HPF**

# IgG4- TIN



- Average age 65 years
- 70-80 % males
- 75 % present with renal disease
- 25% present with renal mass
- 80% have other organs involved \*

**\*Panceas, liver,salivary, lacrimal glands**

# IgG4- TIN



## Laboratory Findings

- **90% have elevated IgG4**
- **60-80 % hypocomplementaemia**
- **30-50 % peripheral eosinophilia**
- **30 % ANA positive (low titer)**

# IgG4-RD



Mikulicz's syndrome (affecting the salivary and lacrimal glands)  
Küttner's tumor (affecting the submandibular glands)  
Riedel's thyroiditis  
Eosinophilic angiocentric fibrosis (affecting the orbits and upper respiratory tract)  
Multifocal fibrosclerosis (commonly affecting the orbits, thyroid gland, retroperitoneum, mediastinum, and other tissues and organs)  
Inflammatory pseudotumor (affecting the orbits, lungs, kidneys, and other organs)  
Mediastinal fibrosis  
Retroperitoneal fibrosis (Ormond's disease)  
Periaortitis and periarteritis  
Inflammatory aortic aneurysm  
Idiopathic hypocomplementemic tubulointerstitial nephritis with extensive tubulointerstitial deposits



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The Annual Nephrology Unit, IMD,  
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Scientific Day 1. Wednesday April 16th, 2014



Kidney & Other Autoimmune Diseases Prof. hussein Sheashaa April 16th, 2014





**Dr.A.P..J.Abdul Kalam**

Past President of India

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END is not the end, in fact E.N.D. means

If you get NO as an answer, remember N.O. means



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مع كل سودا في الأفعير واقتربا  
فلترنا العيسه في أمن يجلل  
وليصدق الفضل في أربابك طربا

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26-12-2013 / المنصورة

